cyanosis, perhaps another hazard function for mortality in their group of patients might have been overlooked.

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Reply

In response to Mee, there are several issues here; the first is semantic. We are reprimanded for "misquoting" the number of infants in the series of Iyer and Mee (1). We read in their report that "four patients were seen in infancy . . . and underwent initial palliation as an emergency." In his letter (and in the report), Mee states that 28 patients >2 years old entered the staging program. These statements are not in fact incompatible; infancy stops at a child's first birthday. In addition, Mee now offers us the information that four neonates were subjected to initial palliation as an emergency. The neonatal period stops at 1 month of age. Even accepting that we may have misread "four patients" as meaning "only four patients," rereading the original report of Iyer and Mee does not clarify how many patients were >1 year old at the time of their first operation. If we have overinterpreted their written statement, we apologize.

Mee also quotes from another report (2) written with coauthors Watterson, Wilkinson and Karl from the Royal Children's Hospital, Melbourne, concerning the same subject. This report (2) was accepted for publication earlier but was published later than the Iyer and Mee (1) report. It included 28 patients with the same diagnosis (1980 to 1989) defined by the nature of their primary procedure and overlapping with the 58 patients described in the Iyer and Mee report (1979 to 1989). Oddly, cross-referencing between the two reports is minimal and is confined to a quote of a survival figure (the 78% suitability figure also quoted in the present letter), which does not in fact appear in the quoted report. I do not think that we can be criticized for failing to reconcile the numbers appearing in the two reports. Neither report deals with patients not undergoing operation who were seen during the same time frame.

All this said, I doubt that there is any real difference in the arguments formulated in our and their reports, namely, that if the natural history of complex pulmonary atresia is to be modified, then intervention in the first year of life will be necessary.

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Familial Cardiomyopathy Associated With Right Bundle Branch Block, ST Segment Elevation and Sudden Death

I read with great interest the report by Corrado et al. (1) of a case of familial cardiomyopathy associated with right bundle branch block (RBBB) and ST segment elevation. This article is a new study of Patient 4 previously presented by the same group (2). However, comparison of the two descriptions provides further information concerning the pathogenesis of this cardiomyopathy:

1. There is a clear change in the pattern of ST segment elevation between the two reports. This confirms the recent observation of Brugada and Brugada (3) of the intermittent behavior of the syndrome of RBBB with ST segment elevation.

2. Electrocardiographic tracings in the present report show a definite (not rate dependent) PR prolongation compared with those tracings presented in the report by Martini et al. (striking in lead II). Because this young adult patient is not receiving drug therapy and histological analysis shows no inflammatory signs, it is probable that the disease process shows rapid progression. We can deduce that (at least in this case) fibrous tissue formation is a histologic sign of disease progression. Therefore, extension of fibrosis independent of lymphocyte infiltrates seems to play an important role in this particular case.

3. In addition, changes in the QRS complex with the disappearance of the R wave in leads V_1 to V_2 over time (unless due to electrode poor position) may be the result of three basic mechanisms of cardiomyocytes transformation: accelerated physiologic cell necrosis leading to fibrosis (as well as the involvement of the conduction system), adipose replacement as suggested by transitional forms evolving from cardiomyocytes to adipocytes (4). Apoptosis is the third, suggested but not yet proved, mechanism of cell differentiation (5).

Corrado et al. deserve credit for presenting the first unmistakable demonstration of definite structural heart disease presenting with RBBB and intermittent ST segment elevation in the right precordial leads. In addition, there is a convincing explanation for the progressive development of an atrioventricular conduction defect involving the right branch with or without the presence of a parietal block. This demonstrates that despite the intermittent behavior of ST segment elevation, the original description of Brugada and Brugada is not always the result of a functional phenomenon.

Nevertheless, the familial cardiomyopathy observed in this young male patient with a normal left ventricle demonstrating replacement of the right ventricular musculature by fatty tissue, surviving cardiomyocytes bordered by fibrosis, without any sign of inflammation, corresponds to the typical clinical and histologic pattern of a previously described form of familial cardiomyopathy called arrhythmogenic right ventricular dysplasia (6).

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Reply

We appreciate Fontaine's interest in our recent article (1). His comments provide an opportunity to clarify certain aspects concerning the pathophysiology of the syndrome of right bundle branch block (RBBB), right precordial ST segment elevation and sudden death. Our study (1) thoroughly investigated a family affected by the syndrome. As Fontaine points out, the proband's clinical findings were anticipated in a previous report from our group that addressed ventricular fibrillation without apparent heart disease (2). Compared with the previous electrocardiogram (ECG), the one shown in the more recent report was recorded later, during the clinical follow-up period, and showed a more accentuated right precordial ST segment elevation. Of note, it was recorded while the patient was taking beta-antiadrenergic drugs. Differences with time in the ST segment elevation pattern, spontaneous or induced by pharmacologic interventions, have been previously reported in this syndrome (3,4). ST segment elevation is characteristically enhanced by vagal maneuvers or class I antiarrhythmic agents and reduced after beta-adrenergic stimulation. These responses have been explained by postulating a local right ventricular (RV) "depolarized" area, which results in changes of ST segment elevation after the above interventions (4). This is even consistent with our hypothesis that RBBB and ST segment elevation are due to a double RV conduction defect, both "septal" and "parietal," which may be modulated by autonomic influences and antiarrhythmic drugs.

With regard to the structural changes of the RV wall, they resembled those observed in the "fatty pattern" of RV cardiomyopathy, with predominant fatty replacement and without inflammatory infiltrates or significant left ventricular lesions (5). However, the coexistence of a severe and progressive (as suggested by the lengthening of the PR interval in the more recent ECG) disease of the specialized conduction system with RBBB interruption, raises some concerns about the nosography of the condition. As recently reported, the conduction system is substantially spared by the dystrophic process of the RV cardiomyopathy but is affected at the advanced stage of the disease, secondary to the septal involvement (6). In our family, there was evidence that the structural changes of RV myocardium and conducting tissue may have been inherited together in the setting of a "heritable cardiac conduction and myocardial disease" (7). Further studies of linkage analysis and gene mapping are needed to better characterize the genetic background of the syndrome.

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